

Research Article

High Resolution Computed Tomography of Chest in Systemic Sclerosis

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Abstract: Systemic sclerosis is a connective tissue disease of unknown etiology involving skin, lung, gastrointestinal tract and kidneys. The most important cause of mortality in systemic sclerosis is the involvement of lungs in the form of interstitial lung disease (ILD) and pulmonary arterial hypertension (PAH). The incidence of ILD in systemic sclerosis varies from 16-43%. High resolution computed tomography (HRCT) is the most valuable imaging in identifying interstitial lung disease in which Non specific interstitial pneumonia (NSIP) is a common pattern observed. HRCT helps in not only detecting the lung involvement, but also in assessing the treatment response. The aim of the present study is to identify the various lesions that are detected on HRCT.

Keywords: High Resolution Computed Tomography (HRCT), Interstitial Lung Disease (ILD), Systemic sclerosis

INTRODUCTION

Systemic sclerosis is grouped into diffuse and limited types based on the pattern of skin involvement. Systemic sclerosis is characterised by typical skin thickening of extremities and trunk with associated pigmentation. The pulmonary involvement is a serious complication of systemic sclerosis. Idiopathic pulmonary fibrosis is more common in the former type. Besides interstitial lung disease and pulmonary arterial hypertension patients can develop pleural effusion, pulmonary hemorrhage, pneumothorax, restrictive ventilatory defect and bronchogenic carcinoma. The lung involvement presents as progressive exertional dyspnoea, fatigue and cough. The pulmonary function test (PFT) is sensitive method in detecting the lung involvement [1]. The chest radiograph findings are non specific. HRCT scan is highly diagnostic in identifying the interstitial pattern in the early part of disease. The HRCT findings in systemic sclerosis include ground glass opacities, consolidation, fibrosis, small centrilobular nodules, pleural effusion, oesophageal dilatation. The change of fibrosis like honeycombing, intralobular interstitial thickening, traction bronchiolactasis are seen predominantly in subpleural and peripheral location in association with ground glass opacities. Posterior and basal segments of lower lobes are mostly affected. Small nodules with or without honeycombing are also seen in systemic sclerosis [2]. The other findings include diffuse pleural thickening in one third of cases, oesophageal dilatation in 58-80% [3], and enlarged mediastinal lymphnodes in 60% of cases [4]. Schurawitzki *et al.* [5] in their study on

HRCT in systemic sclerosis conclude that HRCT was specific in detecting minimal lung disease. The most frequent pattern of ILD seen in systemic sclerosis is non specific interstitial pneumonia (NSIP), but usual interstitial pneumonia (UIP) also occur frequently [6].

PAH is the leading cause of death in systemic sclerosis which occur at an incidence of 10-16%^[6]. There is an increased incidence of lung malignancies reported in systemic sclerosis.

MATERIALS AND METHODS

A retrospective study of HRCT findings in sixteen biopsy proven cases of systemic sclerosis was taken up for the study. Of the sixteen cases thirteen were females and three were males. The age incidence varied from 20-60 years. The main clinical presentations include tightening of skin associated with pigmentation, shortness of breath, dysphagia and joint pains. Routine laboratory investigations were within normal limits. Chest radiograph findings were mostly nonspecific (Fig. 1). Antinuclear Antibody was positive in 14 out of 16 cases and Anti Scl 70 was positive in 11 out of 16 cases. All the patients underwent 2D echocardiography and only 2 patients showed PAH. HRCT of chest was performed in all the patients by 16 slice MDCT (SOMATOM EMOTION of Siemens limited) keeping the standard protocols (KV 130 mAs 120 SL 1.0mm).

In the present study the HRCT findings include ground glass opacities, reticulations, honeycombing, architectural distortion, interstitial septal thickening,

bronchiectasis, traction bronchiolectasis, oesophageal dilatation and pleural effusion. Of which ground glass opacification is the most common observation noted in 11 out of 16 cases (68.7%). Reticular pattern and architectural distortion were the next important finding. Honeycombing and cystic changes were noted in 7 out of 16 cases (43.7%) (Fig. 2-10).

As per the comparative scoring method about the HRCT abnormalities in ILD as described by Wells [7], we have noted that in our study, the most common finding was grade 4 pattern (reticular pattern > parenchymal opacification). Ooi GC *et al.* [8] described a semi-quantitative method of scoring the abnormal HRCT findings by grading each abnormality by the percentage of disease extent and anatomical regions scored. As per the above scoring system the present study showed that the percentage of the disease extent was highly variable.

Goldinn J.G *et al.* [9] (Scleroderma lung study) made a study on the percentage of the disease extent with relation to the zone of involvement. In the present study the percentage of disease extent was 51-75% with variable score.

Honey combing with associated cysts was one of the prominent findings on HRCT in the present study. Mild to moderate cardiomegaly with pleural effusion was noted in five out of sixteen cases. Pulmonary arteries were dilated in only two out of sixteen cases. Oesophageal dilatation was observed in four out of sixteen cases.

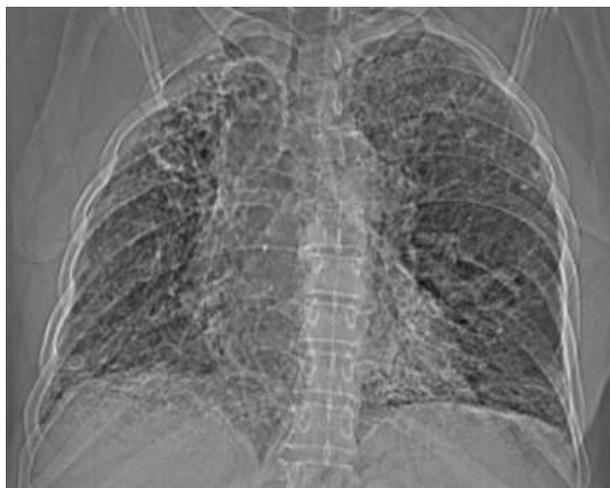


Fig. 1: Chest Radiograph showing diffuse reticulonodular opacities

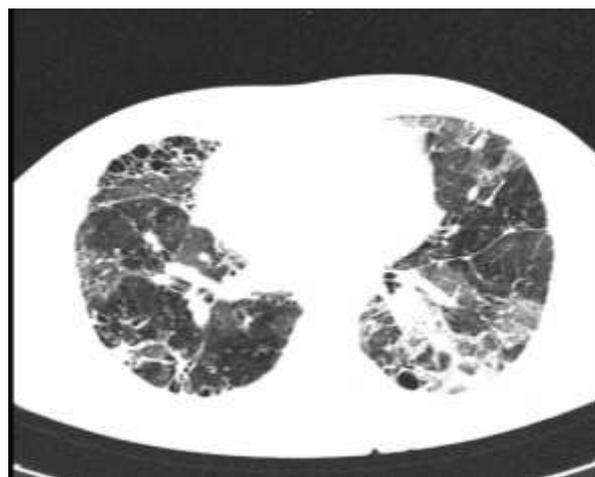


Fig. 2: HRCT image showing patchy ground glass opacification with septal thickening in lung bases



Fig. 3: HRCT image showing bilateral reticulations in upper lobes

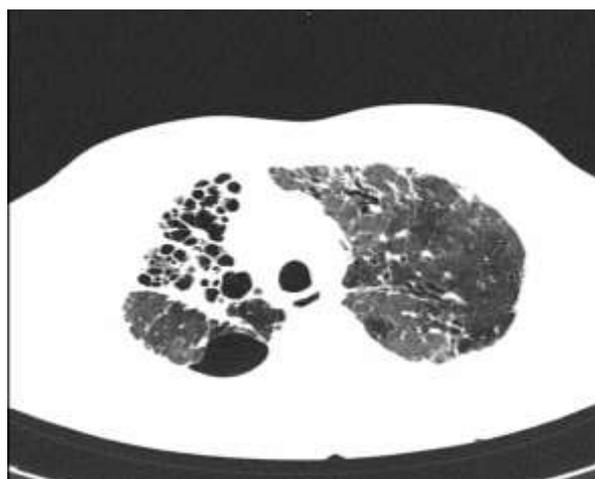


Fig. 4: HRCT image showing honeycombing with cysts and bronchiectasis in upper lobes

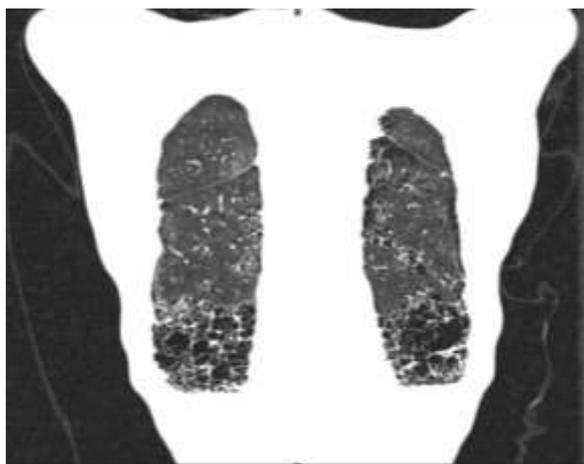


Fig. 5: Coronal HRCT image showing bilateral lower lobe honeycombing



Fig. 8: HRCT image showing dilated oesophagus

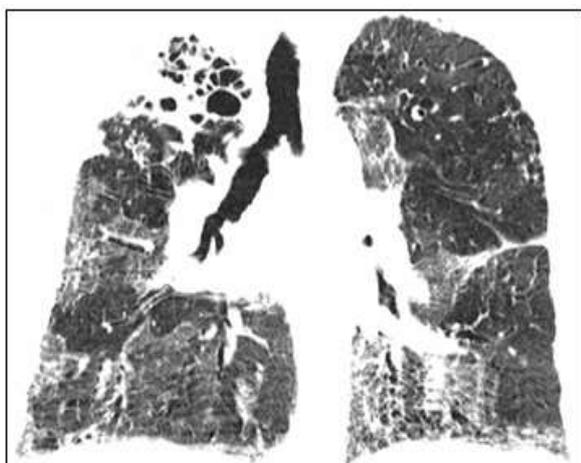


Fig. 6: Coronal HRCT image showing gross architectural distortion of lungs



Fig. 9: CECT Chest showing dilated pulmonary arteries



Fig.7: HRCT image showing cystic bronchiectasis in left upper lobe

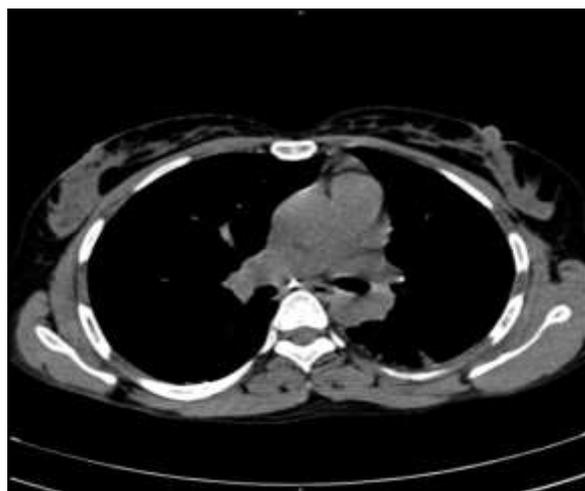


Fig. 10: CT chest showing calcified carinal lymph nodes

DISCUSSION

Pulmonary involvement is the main cause of mortality in patients with Systemic sclerosis [10]. The main types of lung involvement include interstitial septal thickening (ILD), primary arterial hypertension (PAH) and both. ILD is found in upto 90% of the patients with systemic sclerosis at autopsy and 85% by HRCT [1]. HRCT is the gold standard to identify the various patterns of ILD. The major imaging findings on HRCT include ground glass opacities, reticular thickening and both. The presence of ground glass opacities and reticular thickening suggest underlying inflammatory changes, where as predominant reticulations include fibrosis [7]. Ground glass opacities alone is not a reliable indicator of underlying alveolitis and the semi-quantitative evaluation of the disease extent on HRCT is important in evaluating the disease progresss [11]. Different authors attempted to apply scoring systems for the extent of involvement of ILD based on HRCT abnormalities which are classified into comparative, semi-quantitative or quantitative.

In the present study we have observed that ground glass opacities with or without reticulation is the most predominant finding in 11 out of 16 cases (68.7%) followed by architectural distortion and honey combing 9 out of 16 cases (56.2%). Marcel *et al.* [12]. In their study of pulmonary involvement in systemic sclerosis in 23 patients found honeycombing and reticular opacities as most common abnormalities (60.8%).

The semi-quantitative method [13] described the grading of various HRCT abnormalities along with the bronchopulmonary segments involved. In the present study the percentage of bronchopulmonary segments involved cannot be graded, as the abnormalities have no predominant zonal distribution. As per the semiquantitative scoring method described by Ooi GC *et al.* [8] and Scleroderma lung study [9], our study showed the predominant pattern of involvement as mixed ground glass opacities and reticular opacities.

Of the various scoring systems described above, the comparative scoring method [7] is very useful to apply and correlate with the prognosis [14]. The Scleroderma lung study grades the HRCT abnormalities along with the anatomical zones is also reliable in assessing the prognosis of the disease [12].

CONCLUSION

HRCT is highly sensitive in identifying the pulmonary and extrapulmonary abnormalities in patients with systemic sclerosis [15]. Ground glass opacities with reticulations and honeycombing form the most important HRCT findings in systemic sclerosis. Dilatation of oesophagus and the development of pulmonary arterial hypertension aid in the accurate diagnosis of ILD in systemic sclerosis.

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